A LADY WITH HYPERSOMNOLENCE

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30/5/25

- 27y/female
- h/o RTA 13 years ago → Left facial pain → ? left trigeminal neuralgia
- Ganglion blockade in 2020/2022/2024
- Radio ablation of left trigeminal nerve in 2024

- Microvascular decompression on 29th January 2025
- Right eye ptosis on 30/1/25 (POD-1) → resolved spontaneously over 15 days
- Left eye ptosis on 8/2/25 → recovered in 1month

- B/L hearing loss since march 2025

- p/w visual hallucinations / behavioral changes since 14 days
- a/w excessive day time sleepiness
- Bowel/ bladder incontinence
- Imbalance while walking +

NO HISTORY OF :

- Limb weakness
- Sensory complaints
- Seizures
- Fever
- Headache
- Constitutional symptoms

- Admitted in the outside hospital for the same.
- Referred to our hospital for further evaluation

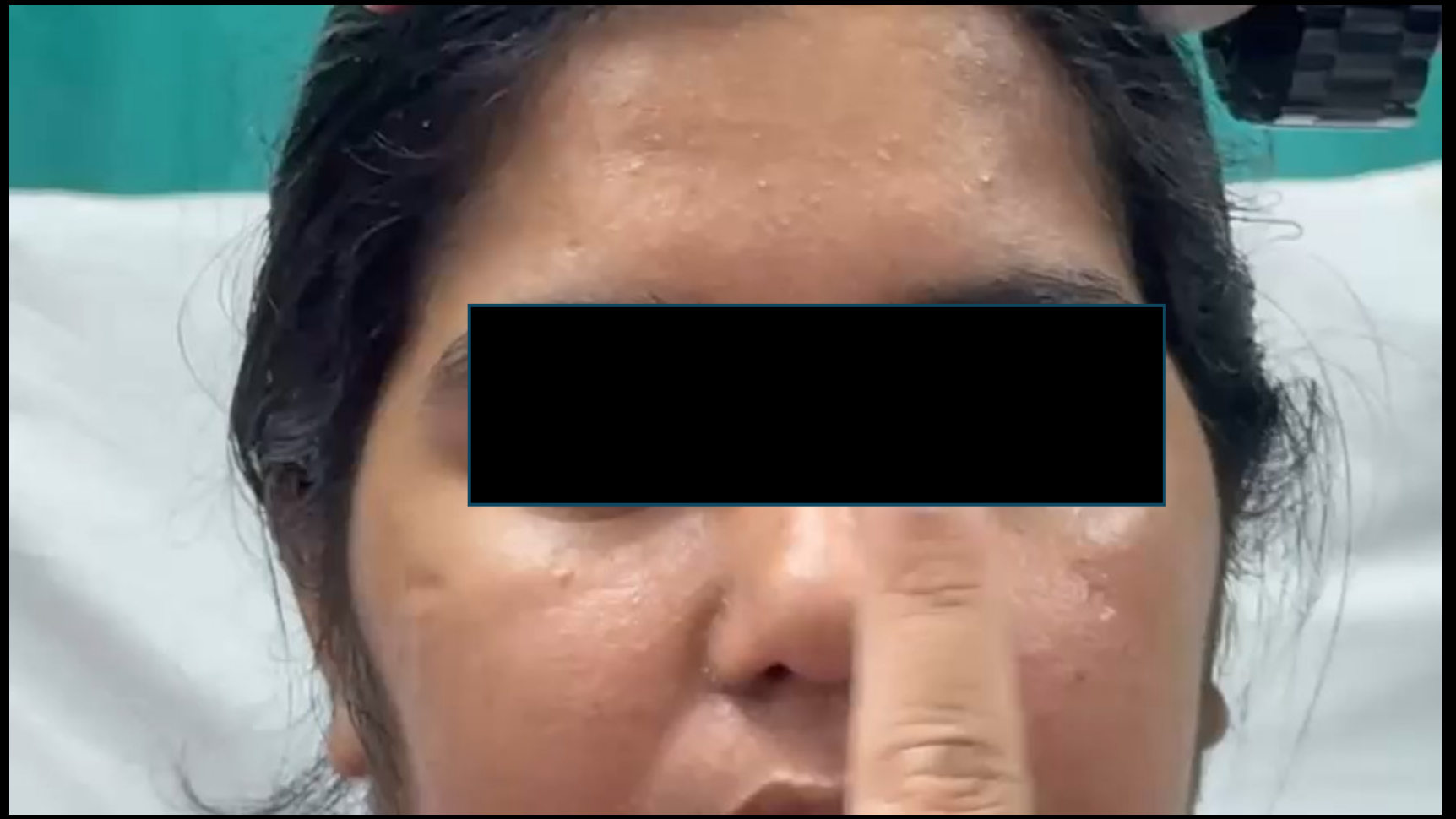
On admission –

- Conscious, following commands
- Disoriented to time / place / person

- Left LMN facial palsy +

- Left eye abduction weakness (lateral rectus palsy +)
- Right eye adduction weakness (medial rectus palsy +)

- B/L gaze evoked nystagmus +
- Finger nose dysmetria + (L>R)
- Dysdiadochokinesia +



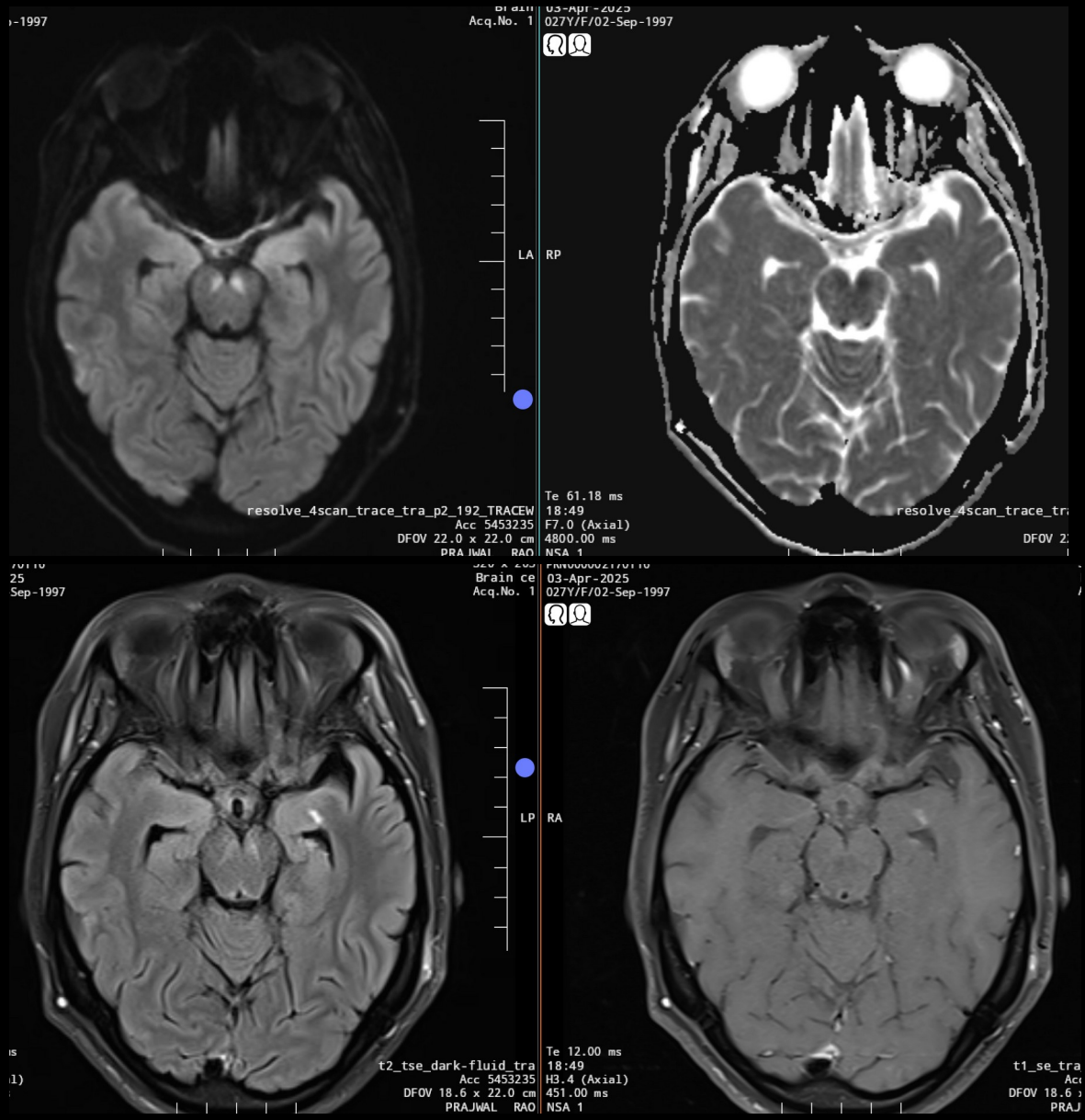
- PR- 88/min
- BP– 110/70 mmHg
- Spo2 – 98% on RA

DIFFERENTIALS :

- ?Brain stem Encephalitis
- ? CNS Demyelination

INVESTIGATIONS :

- Routine labs – WNL
- ANA IFA and Blot- negative
- 2D echo – normal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LA
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DFOV 22.0 x 22.0 cm
PRAJWAL RAO
03-Apr-2025
027Y/F/02-Sep-1997
RP
Te 61.18 ms
18:49
F7.0 (Axial)
4800.00 ms
NSA 1
resolve_4scan_trace_tra
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Sep-1997
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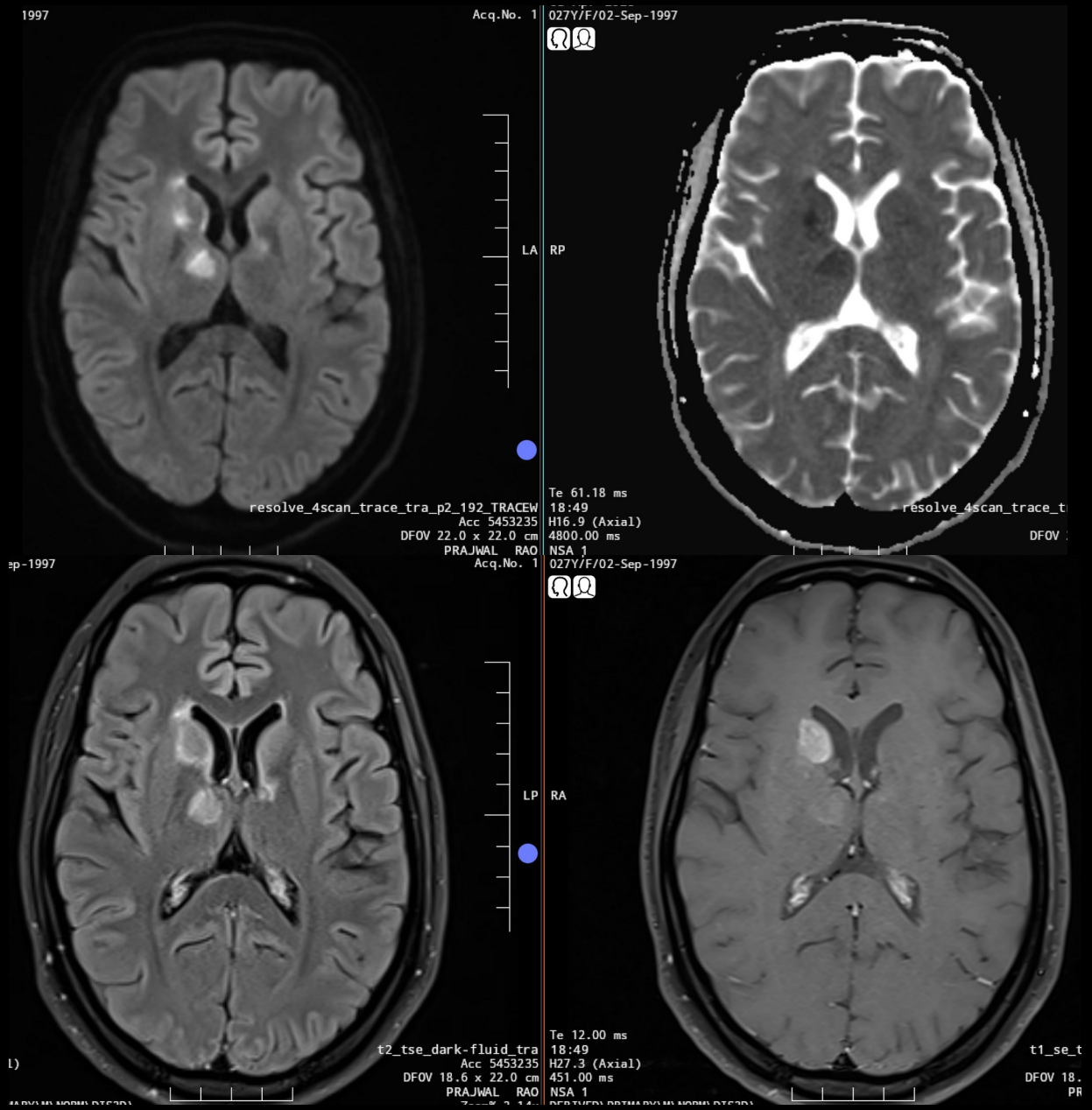
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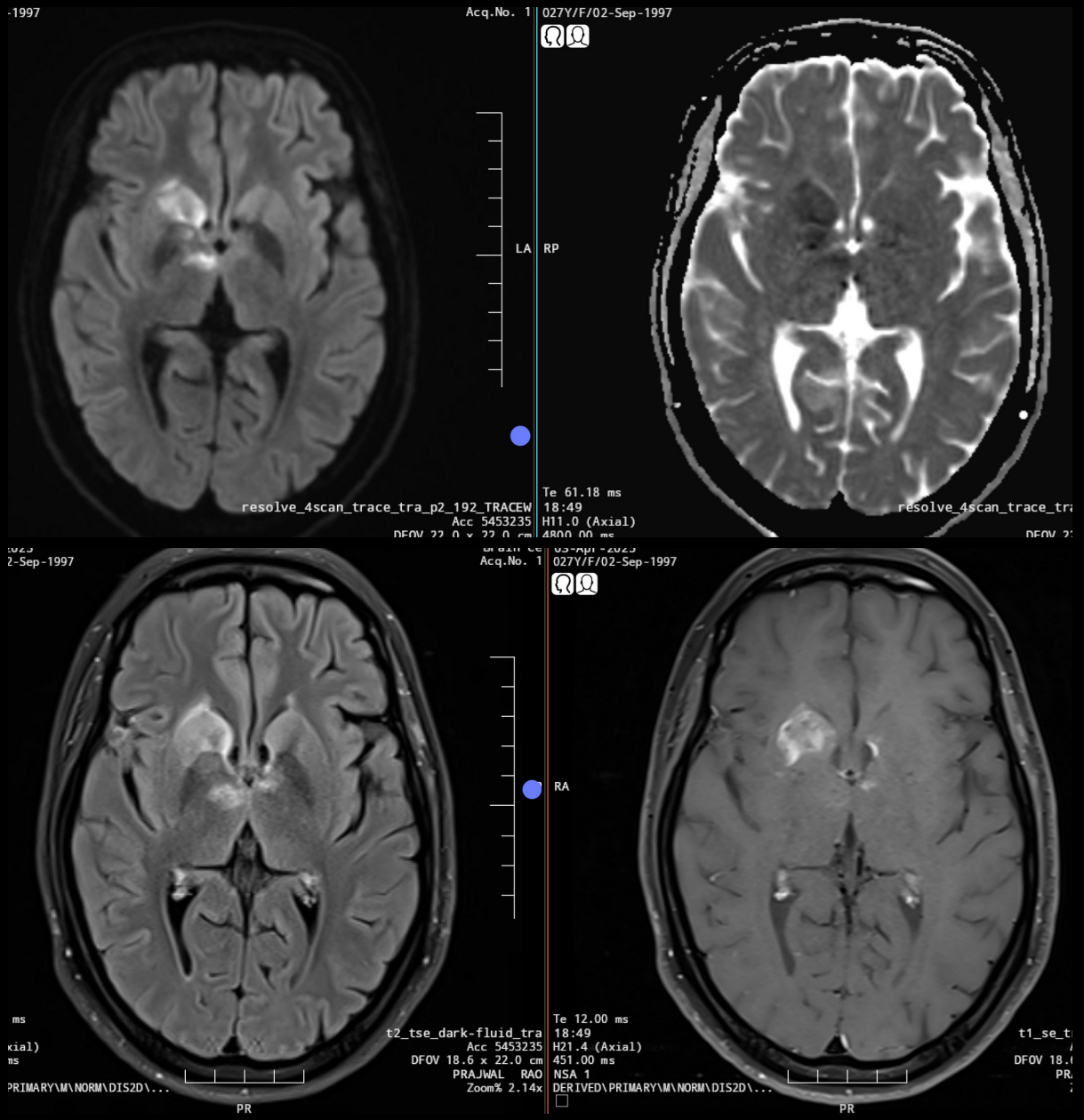
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DFOV 18.6 x 22.0 cm
PRAJWAL RAO
Zoom% 2.14x
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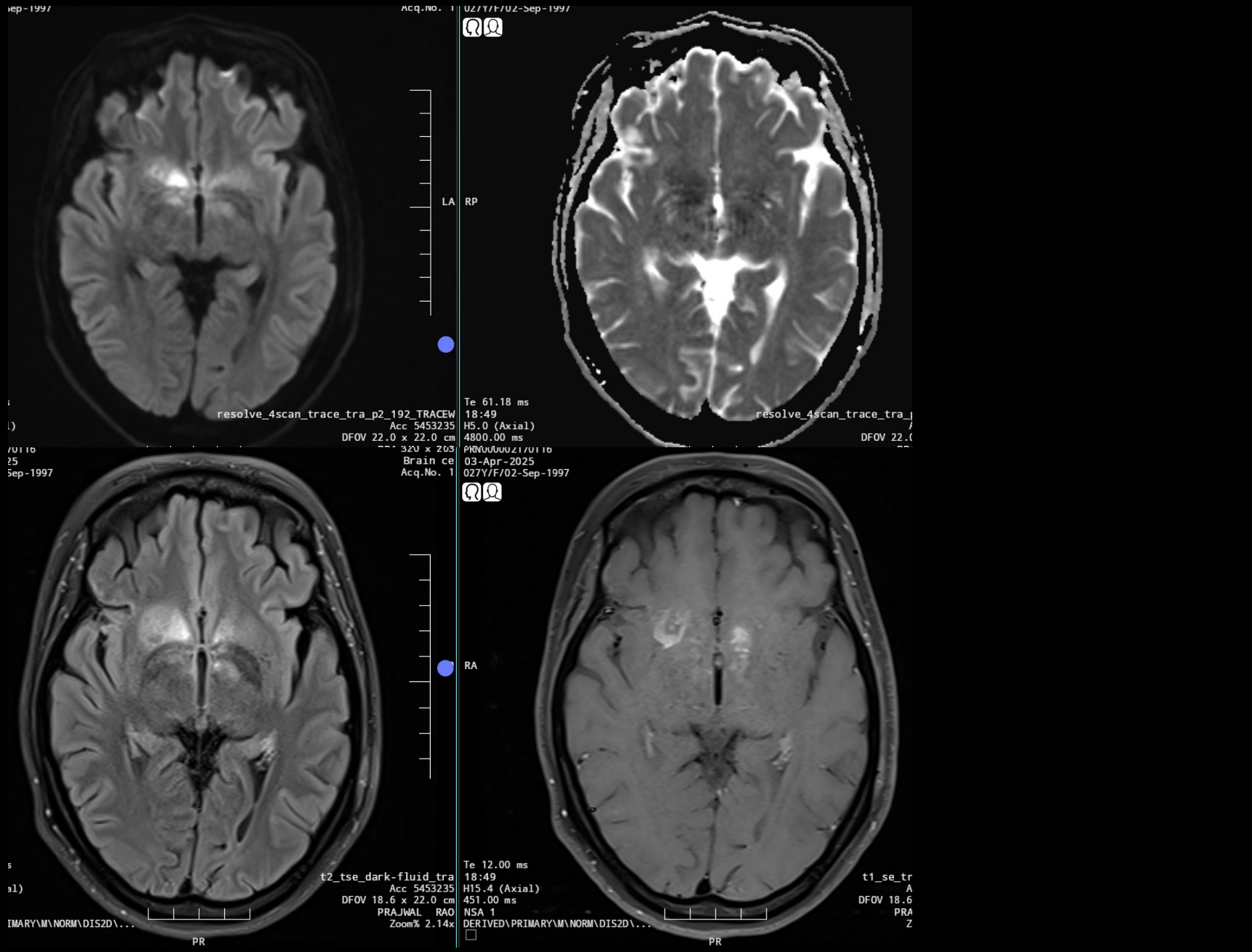
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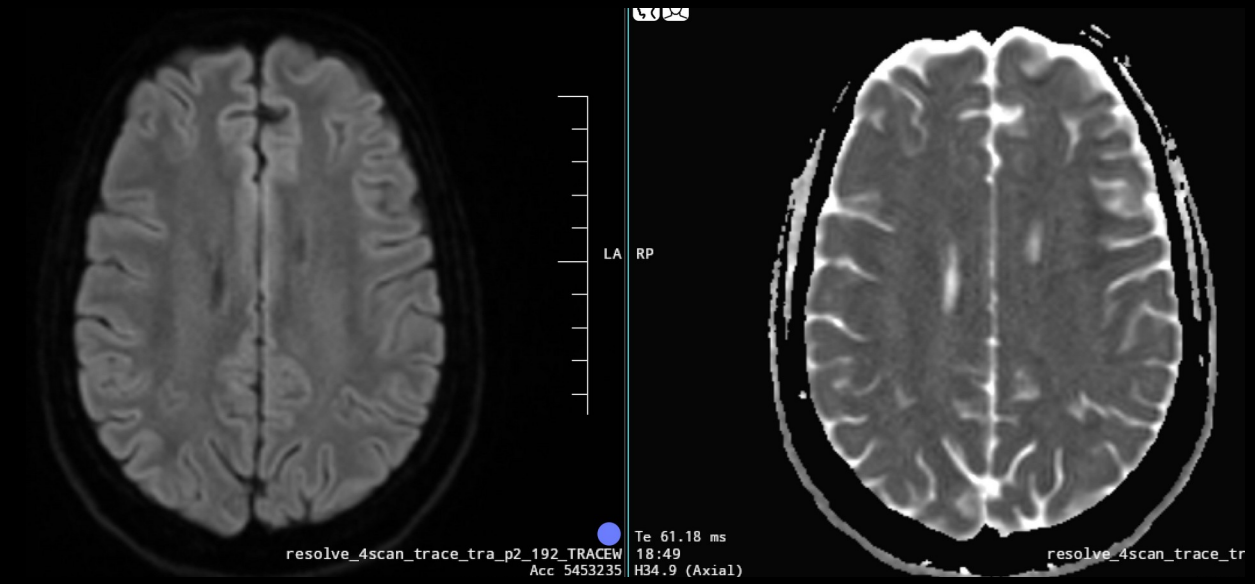
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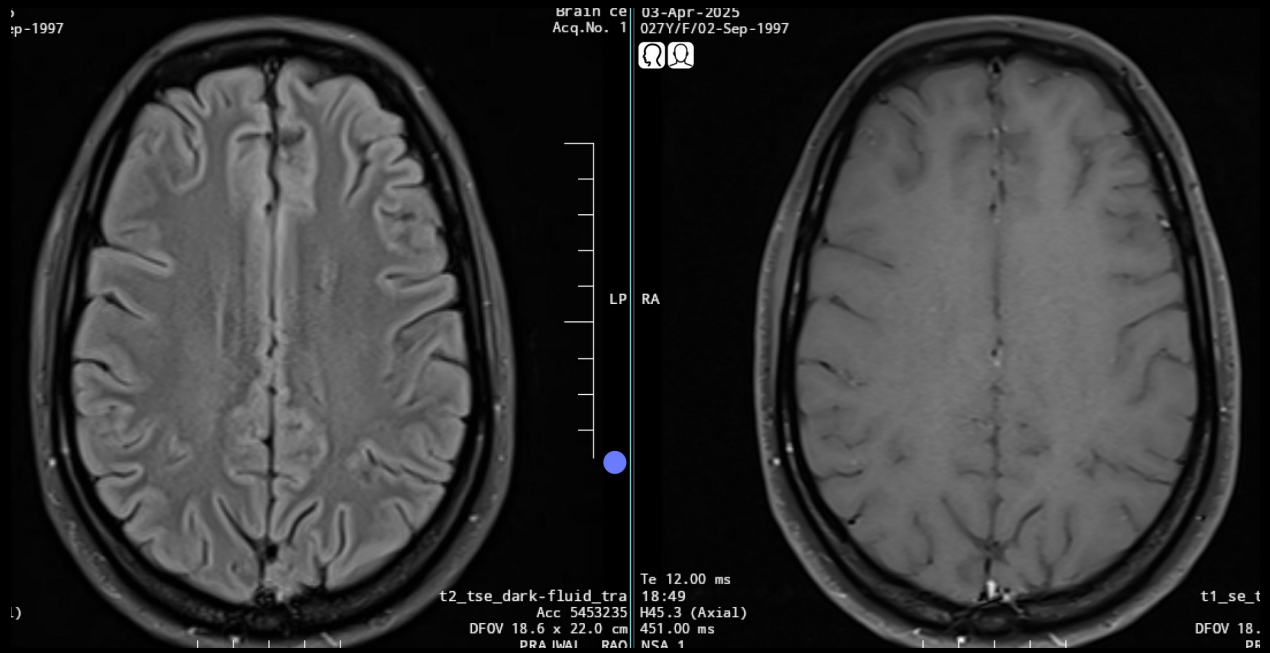
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Acc 5453235
H45.3 (Axial)
DFOV 18.6 x 22.0 cm
451.00 ms
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DFOV 18.

CSF R/M –

- protein – 284
- Glucose – 71
- TLC – 18

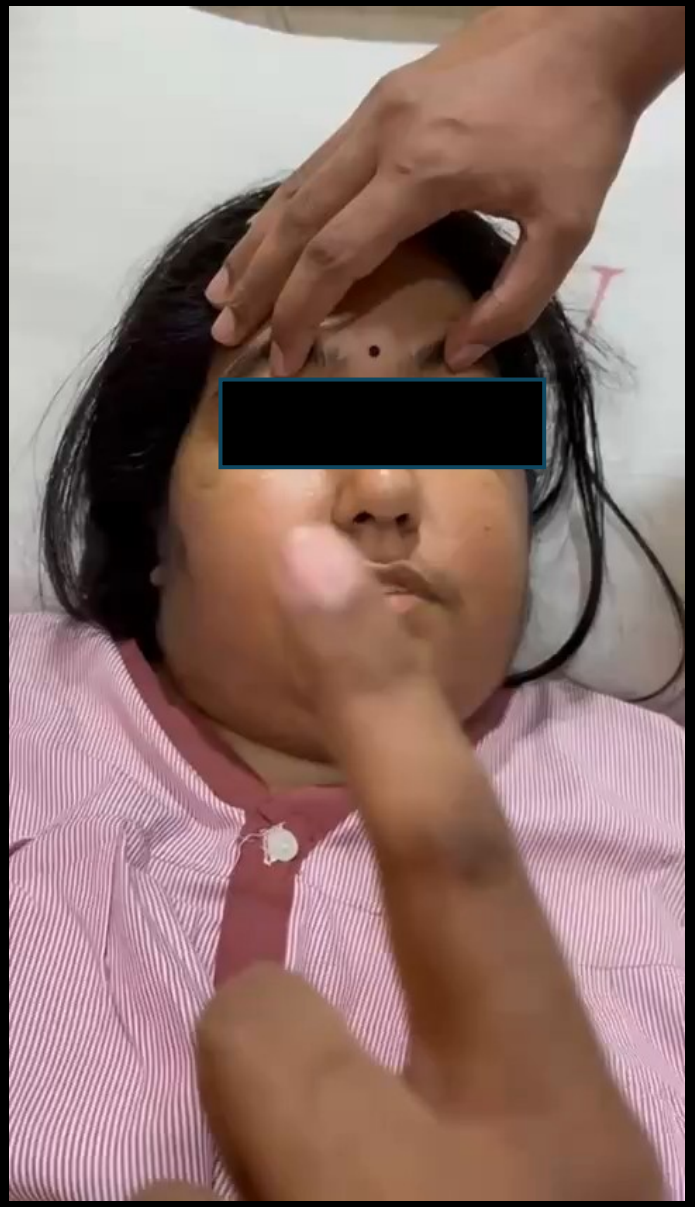
- CSF CBNAAT – NEGATIVE
- CSF CULTURE – NO GROWTH

- NMO – moderate positive
- MOG – negative
- ANCA profile - Negative

- Final Diagnosis – NMOSD-Diencephalic Syndrome

TREATMENT :

- Patient was started on Plasma Exchange
- Completed 7 cycles of plasma exchange
- Started on Inj Rituximab → Currently completed 2 induction doses
- On improving trend i/v/o sensorium, EOMs and gait



DISCUSSION :

- Neuromyelitis Optica (NMO) is a demyelinating disease hallmarked by two distinct presentations: bilateral optic neuritis and transverse myelitis.
- Discovery of Aquaporin-4 antibodies were key in expanding the clinical spectrum of NMO
- Eventually, the term NMOSD was created in 2007.

- Historically, one of the clinical criteria for the diagnosis of NMO was a lack of brain involvement on MRI.
- However, more and more studies have cited the involvement of the brain.
- While brain involvement may seem common, thalamus and sub- thalamus involvement remains relatively quite rare.
- In fact, only 3-4% of NMOSD presents with diencephalic syndrome
- About 1/3rd present with brain stem syndrome.

2015 IPND Neuromyelitis Optica Spectrum Disorder (NMOSD) Diagnostic Criteria

NMOSD With AQP4-IgG

1. At least 1 core clinical characteristic (at right)
2. Positive test for AQP4-IgG*
3. Exclusion of alternative diagnoses**

NMOSD Without AQP4-IgG or Unknown AQP4-IgG Status

1. At least 2 core clinical characteristics (at right) resulting from 1 or more clinical attacks and satisfying all of the following requirements:
   a) At least 1 of: ON, acute myelitis with LETM, or APS
   b) Dissemination in space (≥2 different core characteristics)
   c) MRI requirements, if applicable (at right)
2. Negative test(s) for AQP4-IgG* or testing unavailable
3. Exclusion of alternative diagnoses**

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* Using best available detection method (cell-based assay strongly recommended).
** Evaluation for alternative diagnoses guided by "red flags."

SOURCE: International Panel for Neuromyelitis Optica Diagnosis in affiliation with The Guthy-Jackson Charitable Foundation International Clinical Consortium. www.guthyjacksonfoundation.org/special-projects-and-programs/ipnd-diagnostic-criteria/. Accessed Aug. 24, 2015.

Core Clinical Characteristics of NMOSD

Most common:

1. Optic neuritis (ON)
2. Acute myelitis
3. Area postrema syndrome (APS): episode of otherwise unexplained hiccups or nausea and vomiting

Less common:

4. Acute brain stem syndrome
5. Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions
6. Symptomatic cerebral syndrome with NMOSD-typical brain lesions

Supporting MRI Requirements for NMOSD Without AQP4-IgG

1. **Acute optic neuritis:** brain MRI normal or demonstrating only nonspecific white matter lesions; OR optic nerve MRI with T2-hyperintense lesion or T1-weighted gadolinium-enhancing lesion extending over >1/2 optic nerve length or involving optic chiasm
2. **Acute myelitis:** spinal cord MRI showing attack-associated lesion extending ≥3 contiguous segments (LETM); OR ≥3 contiguous segments of focal cord atrophy in patients with prior history of acute myelitis
3. **Area postrema syndrome:** dorsal medulla/area postrema MRI lesion
4. **Acute brain stem syndrome:** peri-ependymal brain stem lesions

- long corticospinal lesions, hemispheric cerebral white matter lesions and periependymal lesions in the diencephalon, dorsal brainstem and white matter adjacent to lateral ventricles are typical of NMOSD.

- In contrast, juxtacortical, cortical, or lesions perpendicularly oriented to the surface of the lateral ventricle suggests MS as the diagnosis.

Typical NMOSD Brain Lesion Patterns on MRI

1. Diencephalon lesions involving the thalamus and hypothalamus adjacent to third ventricle
2. Cerebellar and dorsal brainstem lesions adjacent to fourth ventricle
3. Dorsal medulla lesions, particularly the area postrema
4. Long, contiguous CST lesions
5. Hemispheric deep or subcortical cerebral white matter lesions
6. Periventricular white matter lesions adjacent to lateral ventricle, including corpus callosum

THANK YOU